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The argument for a non-operative approach to asymptomatic lung lesions

Michael Stanton, MBBS, MD, FRCS (Paed Surg)

Department of Paediatric Surgery, University Hospital Southampton, Tremona Road, Southampton SO16 6YD, UK

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ABSTRACT

The controversy surrounding the management of congenital lung malformations (CLMs) centre on how best to manage the increasing population of asymptomatic antenatally detected infants. Should elective surgery be offered? Or is a "watch-and-wait" policy safe? This will be addressed in this review by examining the reported complications of surgery, the risk of symptom development if lesions are left *in situ* and whether this may alter surgical outcomes, and importantly whether there is any long-term risk of malignancy that can be negated by surgical resection in infancy.

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Introduction

Confirmation that the benign lung lesions of childhood (cystic adenomatoid malformations, bronchopulmonary sequestrations, congenital lobar emphysema, and bronchogenic cysts)[1] could be detected antenatally can be traced back nearly 40 years, to 1975, when Garrett et al.[2] in Sydney, Australia, reported a right-sided microcystic lung lesion associated with foetal hydrops and subsequent stillbirth. Prior to this, cases were identified postnatally with respiratory symptoms or as incidental findings. It is now clear that there is a much larger denominator of asymptomatic cases detected antenatally. Improvements in antenatal sonographic resolution have altered the apparent incidence from initial reports of large symptomatic lesions,[3] through to estimates of 1 in 25,000,[4] 1 in 10,000,[5] up to current estimates of 1 in 2500 pregnancies.[6] In most foetal medicine units, CLMs are likely to be among the most commonly detected anatomical anomaly later seen by paediatric general surgeons. The patient population that clinicians are facing has changed. It is ever more important to understand the natural history of these lesions in order to be able to provide accurate antenatal and postnatal counselling to patients and their families.

Antenatal predictors of poor outcome have been reported, largely related to the volume of the lesion, and may guide the use of foetal interventions.[7,8] A proportion will undergo spontaneous regression or even complete resolution,[9,10] although postnatal imaging is still recommended.[11] Postnatally, most cases are asymptomatic and delivery at a non-tertiary level unit can safely be planned.[11] It is common practice to undertake a plain chest XR on day 1 of life, but to otherwise allow early discharge in the absence of respiratory symptoms. Cross-sectional imaging is usually performed in the first 3 months of life.[11,12] Some neonates (17% in a review) will develop respiratory distress and require early surgery.[10] This is not the subject of debate, and the higher morbidity and mortality reported after neonatal emergency surgery is not unexpected.[13] Elective surgery is advocated in neonates by a small number of centres,[14,15] but surgery is usually deferred until >3 months.[16–18] It is the large proportion of the initial foetal cohort that remains asymptomatic through the first few months of life that are the subject of the controversy. Should they be recommended for elective excision or be managed conservatively?

Will an initially asymptomatic infant with a CLM develop symptoms in infancy?

Symptoms in infants are usually recurrent pneumonia or more rarely pneumothorax. Other general respiratory symptoms such as wheeze/asthma are occasionally reported but are unlikely to occur more frequently in this population. The available evidence suggests that only $<5\%$ will become symptomatic in the first 5 years, although longer-term data are not available yet for large cohorts of conservatively managed infants. A recent systematic review of 41 reports between 1996 and 2008 found that approximately 3% of infants with a CLM *in situ* became symptomatic, at median age of 7 months.[10] This 3% included patients awaiting elective surgery and some in whom expectant management was planned.

In 2014, our centre reported the largest published series of cases in which there was the intention to manage asymptomatic infants conservatively. We found that a similar proportion ($<5\%$)

E-mail address: Michael.stanton@uhs.hns.uk

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became symptomatic after median 5 years follow-up.[19] Of the initial cohort of 74 foetuses, two died *in utero*, one required neonatal lobectomy and three underwent elective surgery as infants at parental request. The remaining 68 infants were asymptomatic at neonatal discharge; three developed symptoms and underwent uncomplicated intervention (lobectomy in 2 and embolisation in 1).

Is the outcome of surgery worse in cases which have become symptomatic before elective surgery?

Proponents of elective surgery argue that the procedure may be more straightforward, and may even have a better outcome, in cases that reported no symptoms before surgery compared to those that had, for example, repeated infections.[11,20,15] This argument has been extended to propose that surgery may be more likely to be completed thoracoscopically in completely asymptomatic cases.[21]

A recent meta-analysis of the outcomes of CLM surgery found that for neonates and infants combined, there was a three times increased risk of adverse events after emergency surgery compared with elective surgery, likely due to the morbidity of neonatal emergency surgery.[10] Complications following elective surgery in infants occurred in 5% (infection, effusion and retained sponge). In the same study, it was not possible to compare the outcome of infant elective and emergency surgery—the group at the heart of the controversy—due to a lack of heterogeneous reports. This last point is worth emphasising, as it is difficult (and probably contentious) to define what being "symptomatic" in the context of an infant with an initially asymptomatic CLM actually means. Some respiratory symptoms occur frequently in the general paediatric population as a whole and it is difficult to know which may be attributable to the CLM. We have defined by hospital admission to treat pneumonia, whereas other reports include symptoms such as asthma, wheeze and upper respiratory tract infections.[15]

Three deaths were reported in symptomatic infants—1 presented postnatally at the age of 8 weeks,[22] 1 underwent pneumonectomy[23] (age not documented) and developed severe pulmonary hypertension and 1 died 5 months after CLM surgery from congenital heart disease.[24] One death (0.3%) was reported following elective resection in an asymptomatic antenatally diagnosed infant.[25]

Thoracoscopy is now widely used to resect CLMs[26,27] and is proving a safe and feasible alternative to open resection. Operative times are longer, length of stay, analgesia requirements and long-term cosmetic result (chest wall deformity) are improved,[21,28] but overall morbidity remains significant.[29] Requirement for open conversion is unpredictable but may be related to the presence of pre-operative infection or final histological diagnosis.[28] Utilisation of the minimally invasive operative approach should not in itself dictate the indication for surgery.

Are there any predictors as to which infants might develop symptoms?

Antenatal features have been reported to correlate with early onset of symptoms in the neonatal period, primarily related to cyst volume.[7,30] Mediastinal shift and polyhydramnios may also be seen, but in our series (and others) this did not correlate with subsequent neonatal symptomatology.[11,19]

There are no prospective studies comparing elective surgery with conservative management for asymptomatic cases. In series reporting symptom development, there are insufficient numbers to determine any lesion-related, or patient-related, characteristics that may predict a higher probability of symptom development. No postnatal radiological markers have been validated to guide whether an infant with a CLM will become symptomatic. Indeed, there is poor correlation with postnatal CT and subsequent histological diagnoses.[31] Logical possible markers would include lesion size and position. Some authors use arbitrary cut-offs to opt for conservative management. For example, Davenport et al.[11] reported leaving small (occupying $>25\%$ of the ipsilateral lung volume), peripheral lesions *in situ*, and Sauvat et al.[12] consider <3 cm as cases where conservative management may be preferable. If one of the proposed intentions of elective surgery is to eliminate the risk of malignancy later in life, then it would be more logical to excise all lesions irrespective of size or anatomical location, in particular as type IV CCAM, with a presumed distal acinar airway origin, may be the lesions having histological overlap with type I PPB.[32,33]

By definition, series describing adult cases currently over the age of 45 years will not have been diagnosed antenatally,[2] and so the difficulty of interpreting against an unknown denominator of asymptomatic undiagnosed cases exists. Several small series exist of adults presenting with lung cysts, diagnosed histologically as CCAM.[34,35] The largest series of adults with CCAM was reported by Makhija et al.[36] where 102 patients, with a median age of 47 years (all >4 years age), were analysed. The frequency distribution was different to that seen in children—bronchogenic cysts was the most common, followed by sequestration, with CCAM occurring in only 8%. Cyst enucleation was performed in two-thirds, with 10% post-operative complications and no cases of neoplasia. This series does provide useful data in the outcome of adults with benign cystic lung disease, but a lack of information about the background group of asymptomatic patients over the same time period, makes it difficult to draw firm conclusions regarding the long-term outcome of current antenatally diagnosed cases left *in situ*. Successful endovascular embolisation has been performed in a fighter pilot allowing continued qualification.[37]

Should CLMs be excised as some may be congenital tumours or undergo "malignant transformation" later in life?

Thorpe-Beeston and Nicolaides stated in 1994 that a decision to operate on all asymptomatic cases "is based on anecdotal evidence of malignant change and is analogous to prophylactic nephrectomy in cases of multicystic renal disease, which has now been abandoned."[3] Twenty years on this remains an interesting comparison. The risk of "malignant transformation," or of a congenital tumour being misdiagnosed as a benign CLM, is regularly quoted to recommend prophylactic lobectomy.[33,38]

The paediatric lung tumours of childhood that have been reported in the context of CLM are pleuropulmonary blastoma (PPB) and bronchio-alveolar carcinoma (BAC).[39,40] Rhabdomyosarcoma (RMS) of the lung is now considered histologically to represent cystic PPB.[41,42] Foetal lung interstitial tumour is a very rare newly proposed entity with only 11 cases reported.[43,44] Most were symptomatic at presentation, and in most cases no additional treatment was required, including in 1 case of known incomplete resection. Primary lung tumours in adults have a median age of diagnosis of 67 years in one recent large series and are strongly associated with smoking.[45]

Bronchio-alveolar carcinoma (BAC) is a very rare lung tumour in childhood, with an overall mean age of 69 years at diagnosis.[46] Five cases had been reported in children up to 1995 (one was a secondary malignancy in a child treated for Hodgkin's lymphoma).[40] BAC has now been reported in association with CLMs in 20 cases, none of whom were diagnosed antenatally with a lung

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